

Rev Mex Neuroci ahora en CONACyT

Vol. 18, núm. 6 (noviembre-diciembre de 2017)

Revista Mexicana de Neurociencia

Publicación oficial de la Academia Mexicana de Neurología A.C.

Revista Mexicana de Neurociencia; 18,6 (2017):96-101

Órgano Oficial de Difusión de la AMN



www.revmexneuroci.com / ISSN 1665-5044

Case report

Gabriel Miranda-Nava,¹Martha
Guadalupe García-Toribio²

¹Clinical Neurologist and
Neurophysiologist, Neurophysiology
department, ABC Medical Center.

²Neurologist and second-year
resident of the subspecialty of Clinical
Neurophysiology, ABC Medical
Center.

Marchiafava-bignami disease: case report and review of the literature

Marchiafava bignami: reporte de un caso y revisión de la literatura

Abstract

INTRODUCTION: Marchiafava-Bignami (MB) is a rare disease characterized by the primary degeneration of the corpus callosum mainly associated with chronic consumption of alcohol. However, this condition can be mistaken for many other diseases which cause lesions of the corpus callosum. Even though it's been said Carducci was the first to report a case of MB in 1898, this disease was named in honor of two Italian pathologists in 1903, Ettore Marchiafava and Amico Bignami, who described the symptoms of an Italian male with excessive consumption of red wine.¹ The majority of patients are male, aged between 40 to 60 years old, with a history of chronic alcoholism and malnutrition. In these cases, morbidity and mortality rates are relatively high. In 2004, there were approximately 250 patients reported, 200 of them died, 30 were diagnosed with severe dementia, and just 20 patients had a positive outcome.²

The treatment for MB is still controversial and shows changing outcomes. Some authors have proposed benefits following the administration of vitamin B, corticosteroids and amantadine.

CASE REPORT: A 41-year-old male with a history of 15 years of chronic alcohol abuse drinking whiskey and rum presents disorientation and minor psychomotor agitation. During physical examination the patient presents dysarthria and ataxy. A family member reports that the patient has presented frequent forgetfulness which has caused him work problems. Nuclear magnetic resonance FLAIR images reveal the presence of demyelination of the corpus callosum, such as a little alteration of the white matter of the brain.

Keywords

Marchiafava, Bignami,
demyelination.

Resumen

INTRODUCCION: Marchiafava-Bignami (MB) es una enfermedad rara caracterizada por degeneración primaria del cuerpo caloso principalmente asociada con consumo crónico de alcohol. Sin embargo, puede ser mimetizada por muchas otras enfermedades que causan lesiones en el cuerpo caloso. Aunque se ha dicho que Carducci fue el primero en reportar un caso de en 1898, esta patología fue originalmente acuñada en honor a los dos patólogos italianos, Ettore Marchiafava y Amico Bignami en 1903, quienes describen los síntomas de un hombre italiano con alto consumo de vino tinto.

La gran mayoría de los pacientes son varones, entre 40 y 60 años de edad, con historia de alcoholismo crónico y desnutrición. En estos casos la morbilidad y mortalidad son relativamente altas. En 2004 aproximadamente se habían descrito 250 pacientes, de los cuales 200 habían muerto, 30 sufrieron demencia severa y sólo 20 pacientes tuvieron resultados positivos. El tratamiento de MB es aún controversial y muestra resultados variables. Algunos autores han propuesto cierto beneficio con el reemplazo con vitamina B, corticoesteroides y amantadina.

REPORTE DE CASO: Se trata de paciente masculino de 41 años de edad que tiene el antecedente de 15 años de ingesta crónica de alcohol en su modalidad de whisky y ron, llegando a la embriaguez de forma constante; se presenta en el servicio con desorientación, agitación psicomotriz; en la exploración cursa con disartria escandida, así como ataxia y alteraciones a la marcha (en estrella). Al interrogatorio indirecto refiere el familiar que ha presentado olvidos frecuentes que incluso le han ocasionado una problemática laboral y en el manejo de dinero. Se realiza estudio clínico que incluye un minimal test, así como el apoyo de neuroimagen a través de resonancia magnética nuclear de encéfalo, donde se muestran imágenes en FLAIR donde resalta la presencia de desmielinización del cuerpo caloso, así como una discreta afección a sustancia blanca contigua, los estudios de laboratorio son normales y no se detecta otra patología agregada, resaltando incluso la normalidad de las pruebas de funcionamiento hepático o electrolitos séricos.

Palabras clave

Marchiafava, Bignami, desmielinización.

Correspondence:

Dr. Gabriel Miranda Nava. Neurólogo y Neurofisiólogo Clínico adscrito al servicio de Neurofisiología del Centro Médico ABC.
E-mail: drgabrielmiranda@hotmail.com

Introduction

Marchiafava-Bignami disease is characterized by demyelination of the corpus callosum. The most frequent clinical features are dementia, attention deficit, walking difficulty, dysarthria, and interhemispheric disconnection syndrome. Pathologically it is characterized by the production of corpus callosum necrosis affecting it locally or totally, and a progressive demyelination of the nearby white matter.²⁻⁴

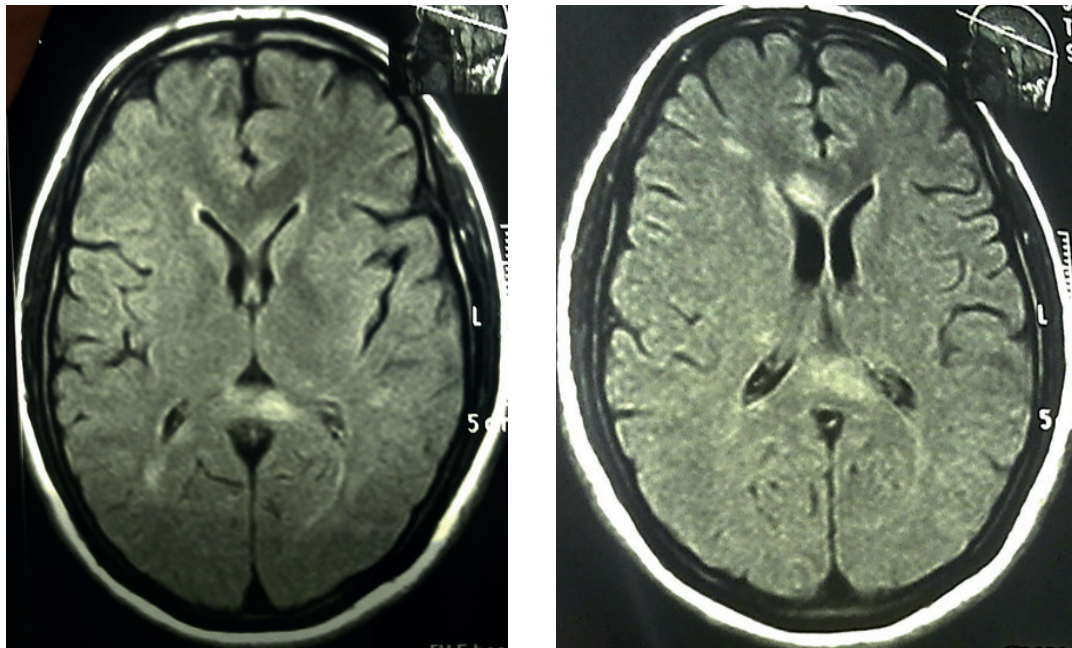
Objetives

The knowledge of this disease as one of many related to the use and abuse of alcohol, as well as its clinical correlation and neuroimaging study.

Clinical case

An interconsultation was requested by the emergency department for a 41-year-old male patient presenting a state of neurological deterioration consisting of chronic disorientation of the three mental spheres, dysarthria, and a constant ataxic or star-shaped gait. An indirect interrogation through his companion reported these symptoms have been present for more than a year. The scan revealed pancerebellar syndrome. The Mini-Mental test scored 18 points out of 30. A nuclear magnetic resonance of the brain was indicated and lesions by demyelination in the corpus callosum were observed. **Figures 1 and 2.**

Figures 1 y2. Nuclear magnetic resonance FLAIR imaging shows the demyelination of the corpus callosum.



Etiology and pathogenesis

The main hypothesis is a deficiency of the vitamin B complex and many patients improve after administration of these components. Other nutritional and toxic etiologies have also been considered and some studies have concluded a racial susceptibility to this disease. Patients without alcohol consumption with MB are mostly younger and are more likely to be female.⁵⁻⁸

Clinical picture

Patients related to chronic alcoholism (often the habit of consumption is a daily and continuous pattern), malnutrition, anorexia, cachexia, or decreased food intake in previous days. Within the clinical manifestations they may present alterations of the mental state (including confusion, delirium, some degree of alertness decrease, memory impairment and/or disorientation), deterioration of gait, dysarthria, mutism, signs or syndromes of disconnection (typical clinical characteristic of the disease), pyramidal signs, primitive reflexes, stiffness, incontinence, sensory symptoms, paralysis of the gaze or diplopia, hemi- or tetraparesis, and nystagmus.⁹

Given the time of clinical evolution, phases have been proposed: acute when the time of evolution is less than two weeks, subacute when it is greater than two weeks, and chronic when it is greater than three months. Diagnosis is based on clinical findings in combination with imaging features. In computed tomography, the corpus callosum appears hypo-attenuated, with the exception of the case that is characterized by subacute bleeding, in which the lesion is shown iso- or hyper-attenuated. During the acute phase, the corpus callosum appears hyperintense in T2 and hypointense in T1. In the subacute phase, the patient may develop cystic lesions and small foci in T2, commonly due to hemosiderin deposits. In chronic stages, alterations of the signal intensity are less evident but a residual atrophy is seen surrounding the structure.^{6,9,10}

Differential diagnosis

We must consider a recurrent artery of Heubner infarct, neoplastic diseases such as astrocytoma or lymphoma, demyelinating diseases such as multiple sclerosis, progressive multifocal leukoencephalopathy, or acute disseminated encephalomyelitis.^{9,10}

Treatment

The etiology of MB is unknown, therefore specific therapy cannot be recommended. Treatment with thiamine and other B vitamins, including vitamin B-12 and folic acid, has shown recovery in some patients; however, therapy failure is not infrequent, even if treatment is initiated at the onset of symptoms. Thiamine has become a treatment option because an association between MB and Wernicke's Disease has been observed in up to 15 to 20% of patients. Patients treated with thiamine in the acute phase have better results than those treated in the chronic phase.^{9,10}

Comorbidities include neuroinfections and epileptic states, among others, which are potentially treatable.

Corticosteroids may decrease cerebral edema, suppress demyelination, stabilize the blood-brain barrier, and reduce inflammation. Some publications have reported cases with improvement after the administration of steroid treatment; despite this, there are other studies in which no net positive effect was observed.

Better results have been observed in patients who are treated with parenteral thiamine within the first two weeks of symptom onset, whereas delayed treatment has been a significant risk factor with poor outcome.⁹⁻¹¹

Follow-up

Acute or subacute MB patients who survive suffer subsequent dementia, though occasionally they may recover partially or completely. Patients who survive the disease should stop drinking alcohol, undergo rehabilitation therapy, and receive nutritional advice. Recovery should be followed by repeated neuropsychological examinations, as well as magnetic resonance control studies, preferably using image diffusion tensor.¹²

Prognosis

A good response is not always observed despite the suggested therapeutics. Some factors that worsen the prognosis are a history of alcoholism and delay initiating thiamine treatment.

The prognosis may also be influenced by the location of the lesion, as solitary splenius lesions are associated with better results compared to lesions located elsewhere or completely crossing the corpus callosum.^{9,10}

Conclusion

MB disease presents a great variety of neurological signs and symptoms. Disconnection syndrome is a cardinal point for the clinical suspicion of this pathology, as well as the relevance of antecedents of alcoholism, malnutrition, and other comorbidities with neuropsychiatric symptoms. These should guide the clinician who, aided by imaging studies, will arrive at a timely diagnosis for the immediate establishment of the therapeutics suggested by the literature, which will give the patient greater opportunities for recovery or at least limit the damage.

Conflict of interest statement

The authors declare there are no relevant conflicts of interest in this study.

Funding

There was no particular source of funding for this scientific report.

References

1. Navarro JF, Noriega S. Enfermedad de Marchiafava Bignami. *Rev Neurol*. 1999; 28: 519-523.
2. Marchiafava E, Bignami A. Sopra una alterazione del corpo calloso osservata in soggetti alcolisti. *Riv Pat Nerv Ment*. 1903; 8:544-9.
3. Gervith RS, Mendizábal RG. Enfermedad de Marchiafava-Bignami. *Arch Neurocién. (Mex)* Vol. 13, No. 2: 138-142; 2008.
4. Yongjian C, Lei Z, Xiaoli W, Weiwen Z et al. Marchiafava-Bignami disease with rare etiology: A case report, *Experimental and Therapeutic Medicine*. 2015; 9: 1515-1517.
5. Samine B, Sirous N, Farrokh ST, Hossein R-D et al. Marchiafava-Bignami and Alcohol Related Acute Polyneuropathy: The Cooccurrence of Two Rare Entities, *Case Reports in Neurological Medicine*. 2016.
6. Sunil K, Rimiki C, Naveen J, Jatishwor S. Marchiafava - Bignami disease: Review of Imaging Features & Case Report, *Journal of Clinical and Diagnostic Research*. 2014; Vol-8(8): RD01-RD02
7. Wenz H, Eisele P, Artemis D, Förster A et al. Acute Marchiafava-Bignami Disease with Extensive Diffusion Restriction and Early Recovery: Case Report and Review of the Literature, *Journal of Neuroimaging*. 2014; Vol 24 No 4.
8. Tekwani PH. Marchiafava-Bignami disease in chronic alcoholic patient. *Radiology Case Reports II*. (2016); 234-237
9. Caulo M, Briganti C, Notturmo F, Committeri G et al. Non-Alcoholic Partially Reversible Marchiafava-Bignami Disease: Review and Relation with Reversible Splenial Lesions A Case Report and Literature Review. *The Neuroradiology Journal*. 2009; 22: 35-40.
10. Hillbom M, Saloheimo P, Fujioka S, Wszolek ZK et al. Diagnosis and management of Marchiafava-Bignami Disease: A review of CT/MRI confirmed cases. *J Neurol Neurosurg Psychiatry*. 2014; 85(2): 168-173.
11. Mestrinelli PE, Marques dos Santos MB, Piasecki L, Cezar JA. Marchiafava-Bignami disease: a rare entity with a poor outcome. *Rev Bras Ter Intensiva*. 2013; 25(1):68-72.
12. Tavares L, Freua F, Kok F. Chronic stage of Marchiafava -Bignami disease. *Images in Neurology*. 2015.
13. Namekawa M, Nakamura Y, Nakano I. Cortical Involvement in Marchiafava-Bignami Disease Can Be a Predictor of a Poor Prognosis: A Case Report and Review of the Literature. 2013; *Intern Med*. 52: 811-813.
14. Augusto L, Figueiredo R, Costa H, Reis C. Marchiafava-Bignami disease as a cause of visual hallucinations. *Rev Bras Psiquiatr*. 2015; 37:82-83.
15. Yadala S, Luo JJ. Marchiafava-Bignami Disease in a Nonalcoholic Diabetic Patient. *Case Reports in Neurological Medicine*. 2013.
16. Furukawa K, Maeshima E, Maeshima S, Ichinose M. Multiple symptoms of higher brain dysfunction caused by Marchiafava-Bignami disease in a patient with dermatomyositis. *Rheumatol Int*. 2011; 31:109-112.
17. Hoshino Y, Ueno Y, Shimura H, Miyamoto N et al. Marchiafava-Bignami disease mimics motor neuron disease: case report. *BMC Neurology*. 2013; 13:208.
18. Rickert CH, Karger B, Varchmin-Schultheiß K, Brinkmann B, Paulus W. Neglect-associated fatal Marchiafava-Bignami disease in a non-alcoholic woman. *Int J Legal Med*. 2001; 115: 90-93.
19. Consoli A, Pirritano D, Bosco D, Postorino P et al. Corticosteroid treatment in a patient with Marchiafava-Bignami disease. *Neurol Sci*. 2014; 35:1143-1145.
20. Kinno R, Yamamoto M, Yamazaki T, Owan Y et al. Cerebral microhemorrhage in Marchiafava-Bignami disease detected by susceptibility-weighted imaging. *Neurol Sci*. 2013; 34:545-548.
21. Heinrich A, Runge U, Khaw AV. Clinicoradiologic subtypes of Marchiafava-Bignami disease. *J Neurol*. 2004; 251 : 1050-1059.
22. Clavier E, Thiebot J, Delangre T, Hannequin D et al. Marchiafava-Bignami disease A case studied by CT and MR imaging. *Neuroradiology*. 1986; 28:376.

Revista Mexicana de Neurociencia, 2017; 18(6): 96-101
www.revmexneuroci.com

Diseño por:

